

CONGENITAL CARDIOLOGY TODAY

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Upcoming Medical Meetings

2016 Pediatric and Adult Congenital Cardiology Review Course

Aug. 21-26, 2016; Dana Point, CA USA
[ce.mayo.edu/cardiovascular-diseases/
content/pediatric-and-adult-congenital-
cardiology-review-course-2016-
general-session](http://ce.mayo.edu/cardiovascular-diseases/content/pediatric-and-adult-congenital-cardiology-review-course-2016-general-session)

Specialty Review in Pediatric Cardiology Course

Sep. 19-23, 2016; Chicago, IL USA
www.pediatriccardiology2016.com/

Sixth Annual Fetal Echocardiography Symposium at UCLA

Oct. 15, 2016; Los Angeles, CA, USA
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Interventional Cardiac MRI

By Kanishka Ratnayaka, MD and Robert J.
Lederman, MD

Interventional cardiologists specializing in Congenital Heart Disease (CHD) have grown adept at using what is available, whether devices or imaging modalities, to treat their patients. Nevertheless, while procedures increase in complexity, operators continue to rely on two-dimensional imaging guidance of gray and white shadows, pattern recognition, and contrast angiography. Complex 3-dimensional spatial relationships are not addressed by current techniques, which can expose patients to significant radiation. Growing and developing children are particularly radiosensitive and carry a lifetime of oncologic risk. Chromosomal damage in the peripheral blood of children exposed to catheterization-related radiation has been detected.^{1,2} Interventional cardiac MRI (ICMR) guidance offers a potential solution.³

Cardiac MRI is a radiation-free, robust imaging modality used to: evaluate cardiac anatomy and function, measure volume and flow, measure tissue infarction, evaluate perfusion and viability, and allow for three-dimensional reconstruction of cardiac and vascular anatomy. Real-time cardiac MRI can provide excellent soft tissue imaging at approximately 5-15 frames/second in many simultaneous planes in any orientation. Combining invasive catheter hemodynamic measurements and MRI physiologic assessment power enables us to realize the full potential of catheterization diagnosis and intervention.

State of the Art

Diagnostic (Invasive)

In patients requiring invasive diagnostic studies, particularly serial studies (single ventricle, heart

transplant) the radiation-sparing argument may be most compelling; the cumulative X-ray dose may be significant.⁴ MRI offers a radiation and contrast-free alternative to those patients who may benefit most from the wealth of structural, functional, and biochemical information MRI can provide. In some critical instances, such as calculating pulmonary vascular resistance in patients with pulmonary artery hypertension and undergoing staged surgical palliation, MRI catheterization evaluation can be superior to the current methods.⁵ While MRI guided catheterization emerged over a decade ago,⁶ it has been non-glamorous, incremental workflow and user interface enhancements that have fueled steady progress. The worldwide experience approaches one-thousand patients. An understandable critique of ICMR is the lack of compatible catheter and guidewire tools, but for invasive diagnostic studies, off-the-shelf balloon endhole wedge catheters are sufficient (Figure 1).

A commercially available MR safe and visible guidewire would enable MRI guidance for most patients requiring diagnostic cardiac catheterization. A polymer guidewire is undergoing final stage clinical testing in Europe,⁷ and safe metallic guidewires are approaching clinical testing.⁸ Another typical critique is that MRI catheterization is time-consuming when compared to current standard X-ray catheterization. In our experience, simple workflow enhancements and experience have substantially decreased time to approximately 15 minutes per hemodynamic condition tested.

The majority of worldwide experience has been performed at three centers (King College London, Great Ormond Street, and National Institutes of Health), but clinical progress has increased attention. Attendance at the Society for Cardiovascular Magnetic Resonance

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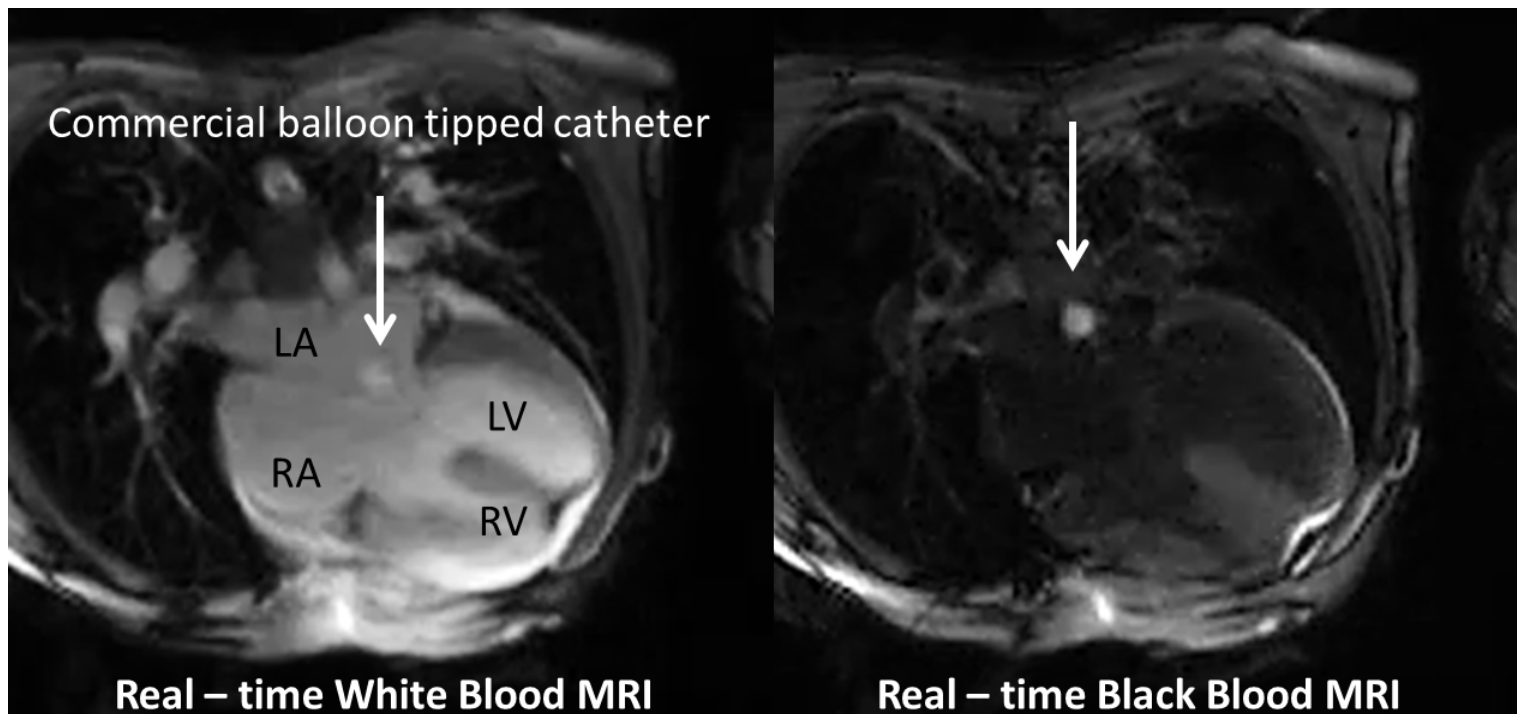


Figure 1. Real-time MRI Right and Left Heart Catheterization in Complete Atrioventricular Canal.

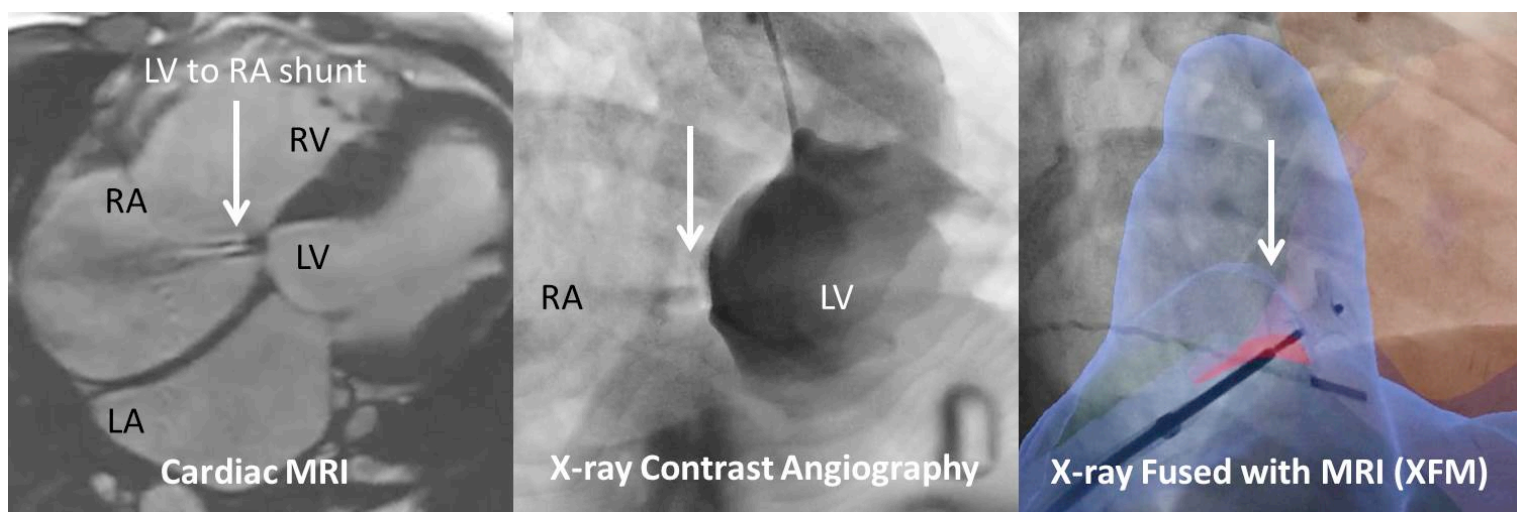


Figure 2. X-ray fused with MRI (XFM) guided device closure of left ventricle to right atrium shunt.

(www.scmr2017.org) annual scientific sessions “interventional cardiac MRI” one day pre-conference has steadily grown with over one hundred participants each of the last three years. In the past year, the National Institutes of Health (NIH) has hosted two hands-on MRI catheterization courses for eighty guests coming from twenty centers in the North America and Europe; future training courses are being scheduled for interested centers.

X-ray Fused with MRI

While MR-guided intervention remains the eventual goal, XFM (X-ray fused with MRI) is an interim step that harnesses the soft tissue information from MRI to guide anatomically and spatially complex procedures. It can be

viewed as a step toward wholly MRI-guided intervention. XFM allows operators to take advantage of the superiority of MRI soft tissue visualization in the familiar working environment of the fluoroscopy suite. The goal of fusion imaging is to enhance the capabilities of X-ray interventional procedures by co-registering MRI-derived roadmaps, to depict soft-tissue features not evident on X-ray. MRI-derived cardiac regions of interest are manually segmented and presented to the operator as image overlay on live X-ray fluoroscopy. Several groups have published on XFM radiation/contrast sparing and enhanced operator confidence in clinical cases.^{9,10} Other groups have shown that registration of static MRI images to live X-ray fluoroscopy takes little time¹¹ with minimal target registration error.¹² Nevertheless, loss

of operator confidence in pre-acquired roadmaps outdated by cardiac and respiratory motion as well as stiff wires and bulky device/delivery systems, continues to be a challenge. XFM may prove most useful in guidance of unconventional interventional Congenital Heart Disease procedures¹³ (Figure 2).

Intervention

Real-time MRI-guided cardiovascular intervention promises superb tissue imaging in multiple views and any orientation to guide traditional and emerging interventional procedures. Pre-clinical MRI guided cardiac intervention has ranged from aortic stenting¹⁴ to aortic endografting to peripheral artery recanalization.¹⁵ MRI guided catheter



Figure 3. Real time MRI guided percutaneous cavopulmonary shunt and transthoracic left atrial access.

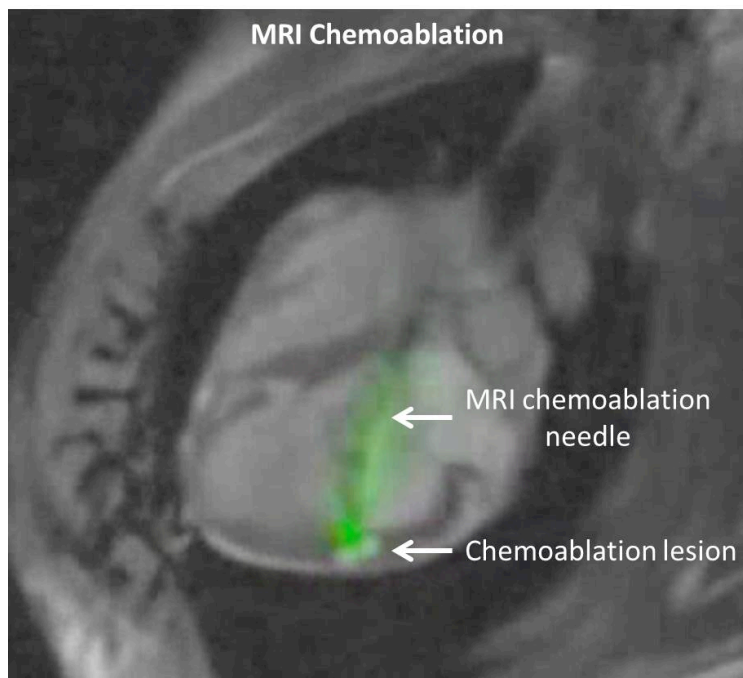


Figure 4. Real time MRI guided chemoablation.

intervention in patients has been limited.^{16,17} Progress in ICMR-guided intervention continues to encounter inadequate MR safe and visible catheter devices. Increasing numbers of small companies focused on delivery of such devices is encouraging.^{7,18}

Interventional cardiac MRI's true potential is in providing surgical-type visualization to enable closed chest, off-bypass novel cardiovascular intervention. One representative example is percutaneous navigation of extra-vascular space under direct (MRI) visualization to join vessels as our surgical colleagues do today with surgical shunts.¹⁹ ICMR provides complete thoracic context imaging that may permit new access routes to the heart for cardiac intervention such as from the patient's back²⁰ (Figure 3).

MR Invasive Electrophysiology

The rationale of MRI guidance for invasive electrophysiology is straight forward - direct observation of myocardial injury during tissue ablation would be attractive to guide procedural conduct; this premise has been explored by a number of groups in animals and most recently in clinical studies.²¹ MRI safe and visible electrophysiology device development has enjoyed tremendous recent progress. An MRI safe and visible integrated catheter mapping and ablation system has been used in clinical translation.²² The device advancement in MRI guided electrophysiology will likely permit significant progression in the coming years. Perhaps more exciting, an alternative approach to tissue ablation using injected caustic agents (acetic acid or ethanol), instead of radiofrequency ablation, exploits the unique capabilities of MRI to map and target arrhythmia substrates and interactively visualize irreversibly necrotic ablation lesions²³ (Figure 4).

MRI Inspired, X-Ray Guided

Cardiac MRI provides operators with a "big picture" view of the entire thoracic context with impressive anatomic detail. Real-time imaging is presented in multiple slices and any orientation that can be manipulated quickly and easily. This ability allows an appreciation of

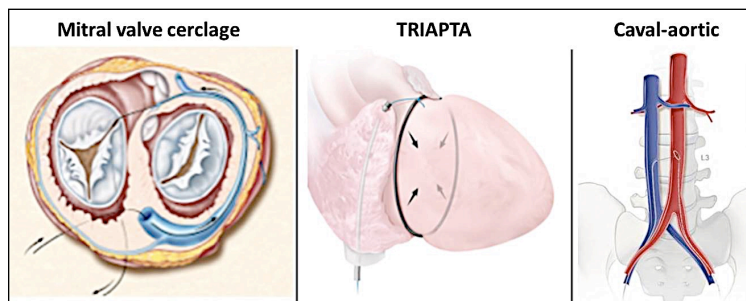


Figure 5. MRI inspired novel percutaneous procedures: mitral valve cerclage, transatrial intrapericardial tricuspid annuloplasty (TRIAPTA), caval-aortic access.

anatomic relationships that is difficult to capture with traditional imaging. Pursuing MRI guided cardiac intervention has inspired innovative X-ray guided procedures. One novel X-ray procedure is percutaneous mitral valve repair by accessing the coronary sinus and tunneling through the myocardium to create a tensioned cerclage loop.²⁴ Exiting the right atrial appendage to deploy a circumferential loop in the pericardium to reduce tricuspid regurgitation is another.²⁵ Exiting the inferior vena cava and entering the aorta to permit vascular entry of large catheter delivery systems and devices is yet another example.²⁶ Clinical translation of caval-aortic access continues to grow. To date, there have been 204 patients at 27 centers (Figure 5).

Conclusions

Minimally invasive and catheter-based therapies are targeting increasingly complex pathologies. This agenda requires better procedural image guidance. Interventional cardiac MRI provides a range of potential radiation-sparing opportunities for conventional and novel therapy.

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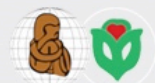


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“Minimally invasive and catheter-based therapies are targeting increasingly complex pathologies. This agenda requires better procedural image guidance. Interventional cardiac MRI provides a range of potential radiation-sparing opportunities for conventional and novel therapy.”

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Biographical Sketch

Dr. Kanishka Ratnayaka is an interventional pediatric cardiologist at Rady Children's Hospital-University of California San Diego. His clinical practice focuses on congenital and structural heart interventions. Dr. Ratnayaka has worked in research collaboration with the National Heart, Lung, and Blood Institute and the National Institutes of Health on interventional cardiovascular MRI for 10 years. Dr. Ratnayaka's other research work includes device development, novel procedures for congenital heart disease, bioresorbable stents for pediatric use.

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Double-Orifice Tricuspid Valve: Case Report with a Review

By DR Hakim Irfan Showkat, MBBCh, MD;
Rekha Mishra, MD; Vinod Sharma, MD, DM;
Lokesh Chandra Gupta, MD, DM; Sadaf
Anwar, PGDCC

Duplication of an atrioventricular valve is an extremely uncommon congenital anomaly that generally affects the mitral rather than the tricuspid valve.¹⁻⁷ The isolated occurrence of this condition seems extremely rare and, in most cases, it is associated with other congenital cardiac malformations that determine a patient's outcome. Even though the Double-Orifice Tricuspid Valve (DOTV) is a rare anomaly and can be easily missed if the physician is unaware of it; it is necessary to scan for an accessory orifice in all patients in whom the atrioventricular valve appears to be small or excessively large.

Case

An 18-year-old girl presented to Outdoor Medical Department with a history of palpitations with mild breathlessness with NYHA Class II-III for a few months. On examination she had a lean and thin build with cardiac examination showing left parasternal heave and normal S1 with split S2. There was a a systolic murmur well heard at apex. Electrocardiography showed sinus tachycardia with right bundle branch block. Two-D Echocardiography revealed Acyanotic Congenital Heart Disease, a Partial-Atrioventricular Canal Defect, a Double-Orifice Tricuspid Valve, gooseneck deformity of LV outflow and a large Primum ASD measuring 4.1 cm with left-to-right shunt. A right atrium and ventricle were markedly dilated with redundant tricuspid valve. There were two orifices of the tricuspid valve with severe regurgitation

“Even though the Double-Orifice Tricuspid Valve (DOTV) is a rare anomaly and can be easily missed if the physician is unaware of it, it is necessary to scan for an accessory orifice in all patients in whom the atrioventricular valve appears to be small or excessively large.”

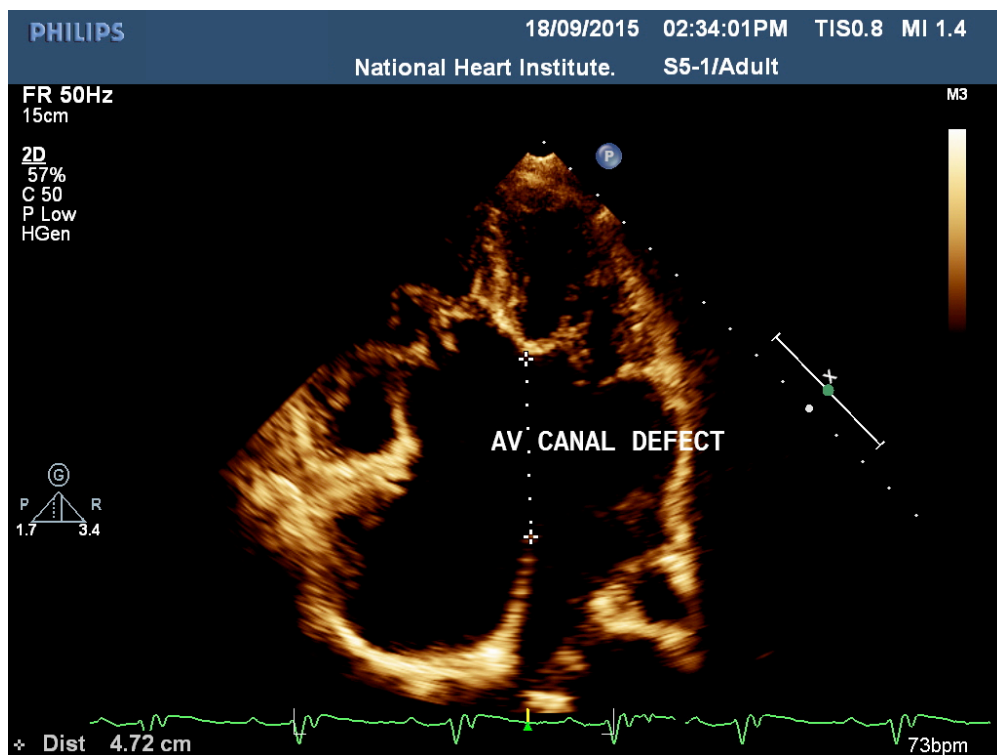


Figure 1.

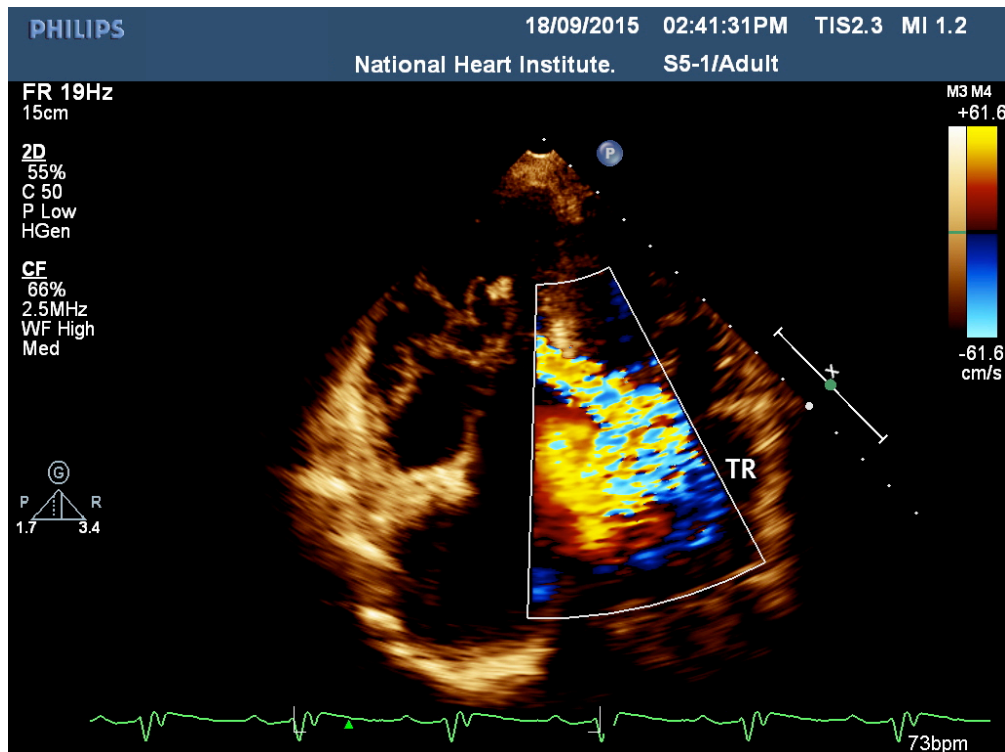
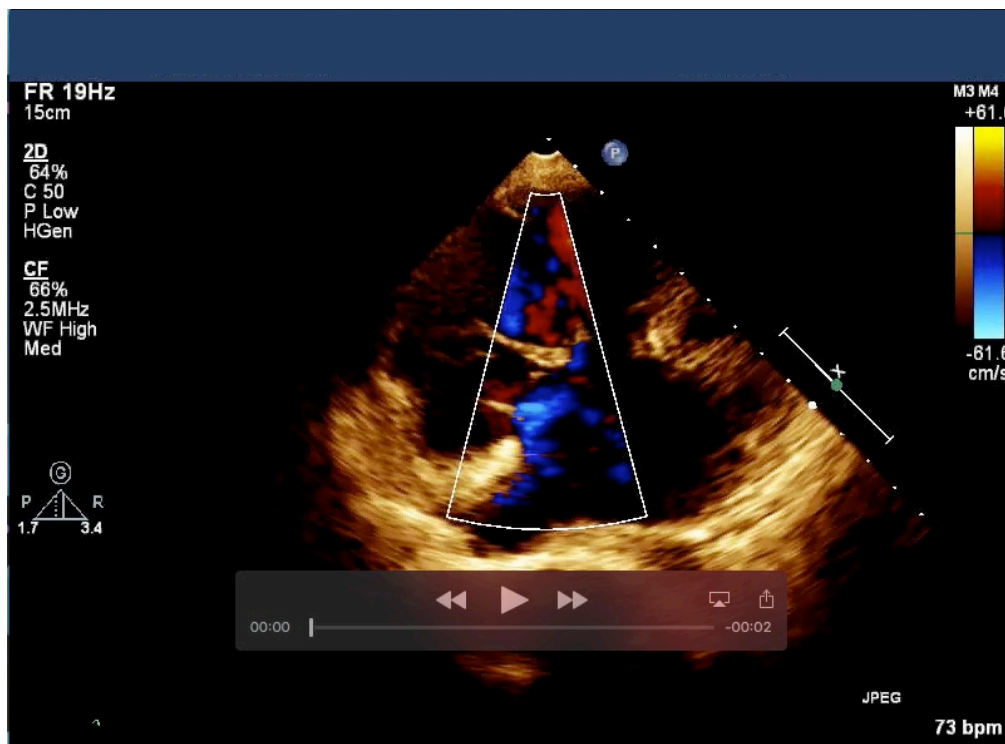


Figure 2.

from one of the valves (valve towards the interventricular septum), low pressure TR jet was eccentric & directed into the left

atrium. (Figures 1, 2; Video 1, 2) Patient was advised to get admitted, but was lost to follow-up.



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Discussion

The division of an atrioventricular valve into two similar and functioning units is described as a duplication of the valvular apparatus or a double-orifice valve. Although this anomaly is well known in the mitral position, duplication of the tricuspid valve is rare. In these situations, it is the presence of an accessory subvalvular component that distinguishes true duplication from a simple fenestration of the valvar leaflet.¹⁻⁷

DOTV is rare and is usually associated with other congenital anomalies, most commonly with septal defects (45%), malformations of the mitral valve, Ebstein Anomaly, and Tetralogy of Fallot.^{3,4,8} To the best of our knowledge, of the 42 reported cases in the literature, only six were isolated.⁴

The first classification of DOTV was given by Hartmann in 1937.¹⁻⁷ The L-type of defect was characterized by two ostia of unequal sizes, the B-type of defect had two equal-sized ostia without an independent set of chordae and a papillary muscle for each ostia and S-type anomalies had two

similar sized ostia and each orifice had an independent set of chordae and a papillary muscle.¹ This classification was revised by Sanchez et al. into three types:

- (1) Commissural-type (Hartmann's type L) in which the accessory orifice is at the end of a valve commissure and its subvalvar apparatus is the normal one for that commissure, though sometimes accessory papillary muscles maybe present;
- (2) Central-type (Hartmann's types B and S) where a fibrous band divides the atrioventricular orifice into either equal or unequal parts as was seen in our case;
- (3) Hole-type, in which the accessory orifice is a hole in a cusp.² This form of double-valve orifice is to be distinguished from a simple fenestration or cleft which has no subvalvar apparatus.

The use of magnetic resonance imaging is helpful in diagnosis as well as its functional significance.⁶

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Dr. Showkat is Editor for *World Journal of Clinical Cases*, and also writes for many other international journals. He has published more than 30 articles and written book chapters on different topics in the field.

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Senior Editor

EchoPixel Medical Virtual Reality System Ready for Clinical Implementation - Breakthrough 3D Imaging Company Offers a New Approach to Surgical Planning and Imaging Diagnostics, Advancing Patient Care with Lifelike Virtual Reality of Patient Anatomy

Marketwire - On July 21st EchoPixel announced that its breakthrough medical imaging solution is now available to clinical users in collaboration with the HP Zvr Interactive Virtual Reality Display and workstation. The HP Zvr, powered by zSpace technology, and the HP Z440 Workstation are customized to EchoPixel's True 3D Viewer cleared regulatory requirements, providing a turnkey solution for both diagnostic imaging and surgical planning.

The True 3D system is a powerful new tool for doctors to make reading medical images more intuitive, help physicians reach their diagnosis, and assist in the planning of complex surgical procedures. The partnership will capitalize on EchoPixel's exciting progress in the study of new clinical applications at prominent beta test sites, and HP's global relationships with medical institutions, to accelerate adoption of

virtual reality technology in the medical imaging field.

Using True 3D, physicians can view and interact with images gathered from CT and MR data the way they would with real physical objects. The system enables radiologists, cardiologists, pediatric cardiologists, and interventional neuroradiologists (among others) to see patient-specific anatomy in an open 3D space.

"I believe our partnership with HP will be a formative moment in the development and distribution of virtual reality in the medical imaging space," said Ron Schilling, CEO of EchoPixel. "HP has a long record of leadership in this industry, a strong network of partnerships, and a powerful commitment to their customers. We believe that virtual reality is the next revolution in medical imaging, and with our FDA cleared system, together we can deliver this technology into hospitals, clinics, and medical schools around the world."

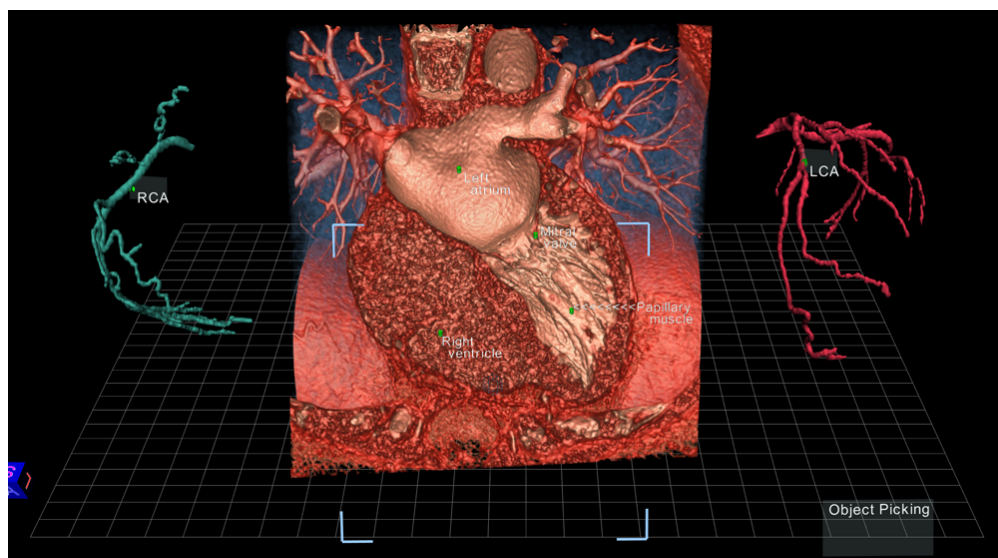
Since its market introduction in March 2015, EchoPixel's True 3D has generated excitement in the medical imaging community, with its promise to transform the ways that doctors work, students learn, and patients understand their unique anatomy. It is being used in clinical, educational, and research settings around the world, including the University of

California, San Francisco, Stanford, the Cleveland Clinic, the Lahey Clinic, and the Hershey Medical Center, among others.

"Our customers rely on HP to help transform lives through innovative solutions," said Reid Oakes, senior director, Worldwide Healthcare, HP Inc. "By working with valued partners like EchoPixel and leveraging emerging technologies like virtual reality, we can rethink how technology can blend the physical and digital worlds to change the face of healthcare."

For further information, or to order EchoPixel True 3D powered by HP, visit www.echopixeltech.com or www.hp.com/go/healthcare.

EchoPixel's FDA-cleared True 3D system uses existing medical image datasets to create virtual reality environments of patient-specific anatomy, allowing physicians to view and dissect images just as they would real, physical objects. The technology aims to make reading medical images more intuitive, help physicians reach diagnosis, and assist in surgical planning. Leading institutions, including Stanford University, the University of California, San Francisco, the Cleveland Clinic, the Lahey Clinic, and more are using True 3D in clinical and research applications.



Coronary CTA. The following is a link to a video showing the EchoPixel in action - <https://vimeo.com/138355486>

First Clinical Use of Bioabsorbable Vascular Grafts in Children Shows Promise

Newsweek — Current cardiovascular valve or blood vessel implants are generally associated with a number of complications, have limited efficacy over time, and may necessitate repeated interventions over a patient's lifetime, especially when implanted in a young child. In a presentation at the 96th AATS Annual Meeting, a team of surgeons from the Bakoulev Center for Cardiovascular Surgery Moscow, report their success with implantation of bioabsorbable vascular grafts used to correct a congenital cardiac malformation. Over time, the grafts are designed to biodegrade as a patient's own cells and proteins reconstitute natural functioning tissue, thus reducing permanent implant-related complications.

Bioabsorbable heart valves or blood vessels are designed to harness the body's innate healing process, enabling the natural

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restoration of complex body parts as the synthetic graft is absorbed. At the 96th AATS Annual Meeting, surgeons from the Bakoulev Center for Cardiovascular Surgery, Moscow, report the results of implantation of bioabsorbable vascular grafts placed into five children born with serious cardiovascular anomalies. According to the investigators, this is the first-ever clinical trial of a bioabsorbable cardiovascular device.

"The positive results of the study provide hope for a new therapeutic approach in cardiovascular valve replacement called Endogenous Tissue Restoration (ETR). This is potentially a revolutionary approach to regenerative medicine in cardiovascular treatment," says lead investigator Leo Bockeria, MD.

The procedure was designed to help children born with single ventricle anomalies, a term used to describe a group of cardiac defects that shares the common feature that only one of two ventricles functions adequately. This can be due to lack of a heart valve, abnormal pumping ability of the heart, or other problems. The surgical procedure, known as a Fontan procedure, involves diverting the venous blood from the right atrium to the pulmonary arteries without passing through the area of the right ventricle.

In this prospective, single-center feasibility study, five children aged 4.5 to 12.5 years born with a single-ventricle congenital malformation were implanted with a bioabsorbable graft connecting the inferior vena cava with the right pulmonary artery during an extracardiac Fontan procedure. Patients were followed for 12 months after surgery using echocardiography, CT-scan and MRI. No device-related adverse events were reported.

The grafts are composed of supramolecular bioabsorbable polymers, manufactured using a proprietary electrospinning process by European medical device company Xeltis. The grafts have no size limitations, although this study used grafts that were 18 and 20 mm in diameter. Histological studies of the grafts in sheep have shown that graft implantation is followed by initial infiltration of inflammatory cells, which induces physiological healing and tissue formation. This is followed by degradation of the

implant scaffold with eventual reduction of the inflammatory response.

The investigators report that all five patients successfully recovered from the procedure, with significant improvement noted in the patients' general condition. Imaging studies demonstrate anatomical and functional stability of the grafts.

Although longer follow-up is needed, the investigators say that the procedure has the potential to improve cardiac and vascular surgical procedures by reducing complications resulting from permanently-placed implants. This is especially important for a child who must live with the after-effects of surgery over his lifetime.

For more information on The American Association for Thoracic Surgery (AATS), visit www.aats.org.

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